

REMARKS ON A CASE OF PORENCEPHALY.

By G. A. GIBSON, M.D., D.Sc., F.R.C.P.Ed., and W. ALDREN
TURNER, M.D., F.R.C.P. Lond.

*Reprinted from the EDINBURGH MEDICAL JOURNAL. Edinburgh and
London, Young J. Pentland, February, 1898.*

REMARKS ON A CASE OF PORENCEPHALY.

REMARKS ON A CASE OF PORENCEPHALY.

By G. A. GIBSON, M.D., D.Sc., F.R.C.P.Ed., *Senior Assistant Physician, Royal Infirmary, Edinburgh*; and W. ALDREN TURNER, M.D., F.R.C.P. Lond., *Physician to the Hospital for Epilepsy and Paralysis, and Assistant Physician, West London Hospital.*

(PLATES IV.—VI.)

AMONGST the pathological conditions giving rise to infantile hemiplegia, those attended by atrophic processes affecting the cerebrum possess deep interest, on account of the difficulties surrounding their causation. Every case belonging to this category is worthy of attention, since it may, even if not giving definite facts of etiological value, aid in the comprehension of the conditions in which it is likely to arise. The circumstances in which the illness of the patient, whose case is about to be recorded, arose, are of value as leading to inferences in regard to the probable causes of the disease, although no real information is to be obtained of a pathological kind.

Clinical features.—J. F., æt. 22, engaged in household occupations at her parents' home, was admitted to the Deaconess Hospital on the recommendation of Dr. Veitch, on 27th January 1895, on account of severe epileptic convulsions. Both of the patient's parents were alive and well, as were also all the members of the family. At the time of her birth they belonged to the well-to-do artisan class, but at the time of the patient's admission to the hospital they had sunk, from various causes which it is unnecessary to specify, into a condition approaching destitution. Apparently the illness for which the patient was admitted, dated from birth. The labour had been a somewhat tedious and very difficult process, and delivery had been effected by instrumental aid, leading to some damage to the patient's head.

Three days after her birth the child is said to have looked nervous, whatever that may mean at such a tender age, and to have been attacked by convulsions, which continued for three days. The motor disturbance was almost entirely confined to the left side. After the convulsions had passed away, the child seemed to progress favourably; but by and by it was observed that, when she began to touch objects, the left hand was paralysed. Her mother had no doubt that the paralytic condition had persisted unnoticed from shortly after the birth of the child. The child appeared to be remarkably healthy, and was rather plump. It was, however, observed that she did not attempt to walk at the usual age; indeed, no progress in that direction was made until she was between 3 and 4 years of age. At this period her mother had her attention directed to a want of symmetry in the shape of the skull.

When somewhere about the age of 7, the patient was sent to school, where she learnt without any great difficulty to read and to spell. Writing, however, she was never able to learn, being only able to copy letters and words when her hand was guided. Her hearing and sight

were always perfect. She had always been of a nervous and excitable nature. It was very easy to produce in her a condition of extreme irritability. Her memory, singularly enough, was retentive, and her speech was entirely unaltered. Shortly after being sent to school she began to suffer from convulsions, which gradually increased in number and frequency.

She left school æt. 14, and since then had always been employed in her parents' house. During the eight years elapsing between her leaving school and the date of her entrance into the Deaconess Hospital, her behaviour was such as might be expected from a child of 7 or 8 years; but immediately after any of her epileptic fits she became somewhat outrageous, and appeared, indeed, to be almost insane. The patient was admitted to the hospital on the evening of 27th January 1895, and, the same night, Dr. Cattnach, then Resident Medical Officer at the hospital, called in order to state that a patient had been admitted in a *status epilepticus*. Thinking that it might be an example of some condition that might be relieved by surgical intervention, we called on Mr. Alexis Thomson, and all proceeded to the hospital together. The patient when seen by us was passing through a series of profound epileptic convulsions, and was absolutely unconscious. The chief peculiarity of the convulsions lay in the fact that each was confined to half of the body, and they occurred approximately in such an order that two convulsions involving the left, were followed by one confined to the right, side of the body, and so on. Each convulsion set in with a tonic spasm of all the muscles of the one side, and at the same time the bilaterally innervated muscles of both sides of the face were thrown into a tonic spasm. The head was invariably turned away from the side on which the convulsions were seen. During the spasm the patient became deeply cyanotic, the face being livid, the lips bluish black, and the veins of the neck turgid. After the lapse of a few seconds after the onset of the tonic spasm, all the muscles which had been affected by rigidity passed into clonic convulsions. During the clonic spasms, the patient threw the arm and leg on the affected side wildly about, and champed with her teeth; but, so far as could be observed, did not damage her tongue. It was impossible, on account of the comatose state and the frequency of the attacks, to make an exhaustive examination of the condition, and no facts could be elicited as to the sensibility and reflexes. The state of the pupils varied considerably, and there were wild movements of the eyeballs. The respiration was much interfered with by the convulsions, and there was involuntary evacuation of the urine. None of this could be collected, so its condition was not ascertained.

Various remedies were tried in order to curb the severity of the attack, but even deep anæsthesia, from chloroform inhalation, had little effect. It was obvious that the patient's end was drawing near, and she died in a state of exhaustion at two o'clock A.M. next morning.

Morbid anatomy.—At the post-mortem examination it was found that the patient was 4 ft. 10 in. in height, quite plump, and well nourished. Muscular rigidity was well marked. Careful measurements were made of the extremities. In length the extremities were exactly equal; but on account of talipes equinovarus of the left foot, measure-

ments were only recorded of the femora and fibulæ, as well as of the humeri and ulnæ. These, thrown into a tabular form, were, in inches :—

	Right.	Left.
Humeri . . .	$12\frac{1}{2}$	$12\frac{1}{2}$
Ulnæ . . .	9	9
Femora . . .	16	16
Fibulæ . . .	13	13

The circumference of the different parts of the extremities were as follows :—

	Right.	Left.
Arm . . .	$9\frac{1}{2}$	$9\frac{1}{4}$
Forearm . . .	$8\frac{1}{4}$	8
Thigh . . .	$17\frac{1}{4}$	17
Leg . . .	9	8

The left half of the head was distinctly larger than the right.

Permission was only granted to examine the brain. On reflecting the scalp, it was very obvious that there was a difference in the size of the two halves of the skull, the left being considerably larger than the right. They measured respectively 10 in. and $7\frac{3}{4}$ in. On removing the calvaria, it was found that the frontal sinuses were very large. The skull itself was somewhat thin. After the brain had been removed, it was seen that the floor of the anterior fossa was somewhat convex, with a deep fissure on each side of the crista. There was comparatively little change in the form of the anterior fossæ, their measurements showing no difference in size. It was otherwise, however, with the middle and posterior fossæ, those of the left side being much larger than those of the right. On measuring the middle fossæ diagonally in their longest dimensions, that of the left side was $2\frac{1}{2}$ in., while that of the right was 2 in. in length. Transversely across, the measurements were $1\frac{3}{4}$ in. on the left, and $1\frac{1}{2}$ in. on the right side. The measurements of the posterior fossæ in the longest diameters showed that the left was $2\frac{1}{2}$ in., while the right was 2 in. The only other point calling for mention was that around the foramen magnum there was a very distinct ridge, with a large protuberance in front towards the left side.

Pathological conditions.—The difference in size and development between the two hemispheres is well seen in the pictures, which are reproduced from drawings by Mr. J. T. Murray. The *right hemisphere*, after hardening in Muller's fluid, weighed $5\frac{3}{4}$ oz. It was partly cystic and partly solid; the cystic portion consisting of the occipital lobe, the convolutions around the posterior end of the fissure of Sylvius, and the ventral part of the temporo-sphenoidal lobe (Plate IV.).

As regards convolutionary development, the gyri over the cystic portion were absolutely atrophied, their place being taken by a dense layer of fibrous tissue, which formed the outer wall of the cavity. The convolutionary areas, which showed the least amount of atrophy, were the gyrus fornicatus, the tip of the temporo-sphenoidal lobe, and the paracentral lobule on the mesial aspect

The remainder of the brain, both mesially and on the convexity, presented a curiously shrivelled appearance (Plate V.), the convolutions being here seen in miniature. The basal ganglia were present, but considerably atrophied. The cavity was lined by a smooth-walled membrane.

It will be seen from this description, and from a consideration of the figures, that although the whole of the right hemisphere is atrophied, that portion which is cystic, and over which the convolutions are entirely absent, marks the distribution of the branches of the posterior cerebral artery; we would, therefore, argue that the primary lesion, probably of a thrombotic nature, occurred in that vessel; and that, although neither the anterior nor middle cerebral arteries entirely escaped, the vascular obstruction was less than that which occurred in the posterior artery.

Examination of the cerebellum and hind brain showed that the *left* cerebellar hemisphere was considerably atrophied in all dimensions. This observation is in harmony with what has been constantly described, namely, a crossed cerebello-cerebral connection, by way of both the superior and middle cerebellar peduncles, that by the superior peduncle being the more direct; as recent observation appears to favour a direct cortical continuation of the superior peduncular fibres. The picture of the base of the brain (Plate VI.) does not bring this fact out. In consequence of the extreme atrophy of the right half of the cerebrum, the cerebellar hemisphere of that side projected upwards to such an extent that its form was altered, and its apparent size, looked at from below, was much smaller than it should have been.

A detailed microscopic investigation of the pons and medulla oblongata was not carried out, but those sections examined showed a partial atrophy of the mesial or chief fillet, on the right side; the portion least implicated being the most internal of the fibres of this structure—the so-called *pes lemnisei*. These sections, therefore, support the view which is now generally accepted, that the mesial fillet ends partly in the optic thalamus, and is partly continued directly onwards to the cortex cerebri; the retention of a considerable degree of fillet formation being in this case accounted for by the presence of the basal ganglia, chiefly the optic thalamus in a fair state of development. There was partial atrophy of the right pyramidal tract.

The brain here figured and described is a good example of the condition called *Porencephaly*, by Heschl,¹ Kundrat,² and Otto,³ in which the cortical defect is in direct association with the lateral ventricle. This is the most advanced pathological condition

¹ *Präger Vrtljschr. f. prakt. Pharmacol.*, 1859 and 1868; *Jahrb. f. Kinderh.*, Leipzig, Bd. xv.; *Arch. d. k. k. Ges. d. Aerzte*, Wien, 1878.

² "Die Porencephalie," Graz, 1882.

³ *Arch. f. Psychiat.*, Berlin, Bd. xvi.

met with in infantile spastic hemiplegia; all degrees being obtained from this downwards through cystic formation, cortical sclerosis, and local softenings, to limited convoluntary atrophy (microgyria).

Various views have been propounded as to the causation of this condition, but that most favourably received, and which fits the case under consideration most nearly, is a thrombotic occlusion of a cerebral artery, with secondary softening and cystic formation. In this case, though the entire cerebral hemisphere is atrophied, it would appear that the posterior cerebral artery was that chiefly implicated. That this may take place before birth seems highly probable in some cases. The history in our case clearly points to the onset three days after birth, when the child was seized by convulsions; and the conditions in which the first clinical appearances were developed seem in favour of thrombosis rather than of embolism as the cause of such vascular occlusion, as was first seen by Heubner.¹

The subsequent onset of epilepsy in such cases is invariable.

¹ *Wien. med. Bl.*, 1883.











